Radiology Corner

Schistosomiasis Japonicum Involving the Liver and Colon

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Note: This is the full text version of the radiology corner question published in the January 2007 issue, with the abbreviated answer in the February 2007 issue.

This case report reviews the general characteristics and life cycle, clinical presentation, typical imaging findings, and treatment of Schistosomiasis. Schistosomiasis is a chronic infection caused by parasitic trematode worms that currently affects 200 million people in subtropical and tropical environments and is not an insignificant threat to deployed military members. (1) Schistosomiasis infection in humans begins with cercariae penetration of the skin or buccal mucosal from a contaminated water source. Schistosome ova are subsequently deposited within the liver, gastrointestinal tract, and genitourinary tract and a granulomatous and fibrotic reaction ensues, resulting in a multisystemic nonspecific and often clinical manifestations. Schistosomiasis is major cause of portal hypertension worldwide and can potentially result in permanent gastrointestinal and urinary system damage if not appropriately treated.

History

A 51-year-old female presents with chronic left flank and pelvic pain. Her past medical history is remarkable for a remote liver biopsy showing Schistosomiasis japonicum infection. She reports prior travel to several Asian countries. Her past surgical history is notable only for a prior cesarean section. The physical examination shows a nondistended abdomen and nonspecific lower abdominal pain and was otherwise noncontributory. Abdominal radiography (Fig 1) and CT of the abdomen and pelvis (Figs 2a-d) were obtained.

Summary of Imaging Findings

Imaging is useful for showing the extent of involvement and identifying complications of disease. Definitive diagnosis is achieved with serologic testing or tissue biopsy. Drug therapy is the primary form of treatment and can lead to rapid clinical improvement.

Abdominal radiography showed subtle curvilinear densities overlying in the left lower quadrant and central within the pelvis (Figs. 1a,b). This was confirmed on intravenous contrast-enhanced abdominopelvic CT to be the caused by thin mucosal surface calcifications within the descending and sigmoid colons (Figs. 2a,b). Incidental note is made of thin right hepatic lobe capsule calcifications (Figs. 2c,d). Bladder calcification was not present on remainder of pelvis CT (images not included). These colonic and hepatic calcifications are characteristic of Schistosomiasis infection of the gastrointestinal tract. Liver biopsy confirmed Schistosomiasis japonicum infection.



Figure 1 Fig. 1a. Anteroposterior abdominal radiograph shows subtle linear densities in the pelvis and left lower quadrant (arrows).

Discussion

Schistosomiasis (aka bilharzias or "snail fever"), is a chronic parasitic illness that, although rare in the United

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Form Approved OMB No. 0704-0188 States, affects between 200 to 300 million people across the world (1), with another 600 million people at risk of contracting the infection. This disease is endemic to 74 countries to include Central and South America, Egypt and other African nations, the Middle East, Asia (particularly rural areas in China), and India. The infection is brought to the United States by immigrants and travelers returning from endemic areas.



Fig. 1b. Magnified AP abdominal radiographic image shows that these densities may be thin linear calcifications that have a tram track-like appearance (arrows).

The main species of blood flukes or Schistosomes to infect humans are S. mansoni, S. japonicum, S. mekongi, and S. haematobium. The first three species characteristically affect the gastrointestinal tract (inhabiting the portal veins) and the latter typically affects the urinary tract (inhabit veins of bladder) and is the most common cause of bladder calcification in the world. The incidence in males is nine times greater than in females. Risk factors include but are not limited to extreme poverty, lack of public health facilities, and poor sanitary conditions. Infection most commonly occurs in adults who live in rural areas and/or who work in either the agricultural or freshwater fishing fields.



Fig. 2a. Axial intravenous contrast-enhanced CT image shows thin mucosal calcification (arrow) along a length of the descending colon, corresponding to the radiographically-identified linear densities within the left lower quadrant.

The Schistosome has a complex life cycle: humans are infected through contact with contaminated fresh water. The pathophysiology of gastrointestinal calcification is related to the Schistosome fluke eggs deposited in tissues and not caused by the flukes themselves. Following deposition of these Schistoma fluke eggs within the venules of the urinary tract (most commonly the bladder and distal ureters), and gastrointestinal tract lumen, there is development of a hypersensitivity or granulomatous reaction and fibrosis around the Schistosome eggs. The severity or degree of calcification correlates with the number of eggs deposited. Many of the eggs are swept back into the liver, where they become lodged within the parenchyma and result in hepatic calcification, fibrosis and associated portal hypertension.

Clinical manifestations can be separated into three different phases: acute, chronic, and severe. Also known as Katayama fever, the acute phase is usually found in children or young adults who have not had any prior exposure to Schistosomes. Believed to be the result of high antigen exposure, this early phase is more commonly associated with S. japonicum infections. Clinical manifestations of this early phase include: bronchospasm, fever, dermatitis, malaise, lymphadenopathy, and arthralgia; amongst other nonspecific complaints. The chronic phase, which is usually subclinical, is characterized clinically by diarrhea and fevers. There may be related growth rate depression, hepato- and/or splenomegaly, and complications of hepatic fibrosis (i.e. portal hypertension, hepatosplenomegaly, ascites, varices of the esophagus and related hemetemesis).

The most frequent cause of death in hepatosplenic schistosomiasis is gastrointestinal bleeding (2). Bladder fibrosis may contribute to ureteral obstruction, pyelonephritis, and hydronephrosis which can lead to renal failure. Bladder infection has been linked with the development of bladder

carcinoma. The severe phase of disease, unlike the chronic phase, is typically symptomatic.



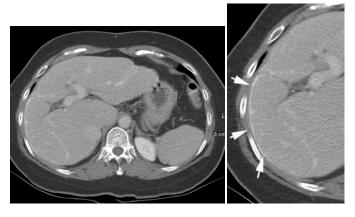
Fig. 2b. Axial intravenous contrast-enhanced CT at a lower level in the pelvis shows similar tram-track calcification pattern of the mucosal surface of the sigmoid colon (arrows).

The diagnosis can be readily made through the identification of eggs in stool in S. japonicum and S. mansoni infection or by detecting eggs in urine in S. haematobium infection. Other more aggressive means of definitive diagnosis include liver, rectal, or bladder biopsy. Additonal nonaggressive yet relatively sensitive diagnostic tests include oval precipitaiton testing and an intradermal immunological skin test for the Schistosome antigen.

Imaging can play a role in demonstrating the extent of organ involvement and complications of Schistosomiasis infection. Conventional radiography may be the initial means of detecting calcification of the bladder or distal ureters (parallel linear radiodense appearance). Intravenous pyleogram (IVP) or cystography in particular are very good studies for demonstrating ureteritis cystica, ureteral stricture (or dilation), hydronephrosis, mucosal irregularity, urolithiasis, decreased bladder capacity, bladder tumor, and inflammatory pseudoployps. CT is useful and more sensitive than radiography for the detection and characterization of urolithiasis and pyelonephritis.

Nodular hepatic lesions have been demonstrated sonographically and by CT in acute Schistosomiasis (3). In the colon, extensive curvilinear or tram-track calcifications have been identified in patients with schistosomiasis japonica (4). Another study involving intravenous contrast-enhanced CT in Schistosomiasis japonica hepatic infection, showed septal, amorphous, and capsular contrast enhancement patterns. This study concluded that septal enhancement may suggest the diagnosis of hepatic schistosomiasis japonica, particularly when calcification is not identified on noncontrast CT study (5). The most reliable diagnostic techniques for

detecting chronic bladder disease are cystoscopy, serology, CT and to a lesser extent urography. Urinalysis in chronic disease may result in missed detection of disease that would otherwise be identified during the early phase of disease (6).



Figs. 2c,d. Axial CT image of the liver and magnified image of the right hepatic lobe shows thin hepatic capsular calcifications (arrows). Note also the prominence of the portal vein, the slightly shrunken overall appearance of the liver, and the mildly nodular hepatic surface, findings indicative of chronic liver disease.

Differential considerations for bladder calcification (when present) include: Tuberculosis (TB), alkaline incrustation cystitis, primary amyloidosis, and radiation. While TB typically descends from the kidneys to involve the bladder, Schistosomiasis behaves in the opposite manner, beginning in the bladder and ascending to involving the kidneys at a later stage. Despite involvement by fibrosis, the bladder remains distensible.

The primary treatment of schistosomiasis is the use of drug therapy, namely praziquantel, oxamniquine, and metrifonate. Early treatment can lead to rapid clinical improvement. Extensive fibrosis is associated with irreversible disease. Bleeding esophageal varices can be managed by sclerotherapy or surgery. As of yet, there remains no vaccination for this disease; however, research is currently being conducted towards producing a safe and effective protective vaccine as a long-term solution to this significant health problem (7).

Note: Follow this link for Category 1 CME or CNE in the case of the week in the MedPi \mathbf{X}^{TM} digital teaching file.

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